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An atypical presentation of retroperitoneal fibrosis as an adrenal mass: diagnostic challenges

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A 58-year-old man without prior medical history has presented to his general practitioner with nonspecific abdominal discomfort. The patient denied trauma or weight loss. Laboratory tests, apart from increased total cholesterol, gamma-glutamyl transferase and platelet count, show no abnormalities. Inflammatory markers were negative (the C-reactive protein level was 4.5 mg/L). Abdominal ultrasound showed an abnormal mass in the left adrenal area, further characterized on magnetic resonance imaging (MRI) as an irregular 85 x 62 x 72 mm tumor with central necrosis, adjacent to the left diaphragmatic branch of the fundus walls and body of the stomach, spleen, kidney, lumbar muscle, modelling the visceral trunk and superior

mesenteric artery. Abdominal computed tomography (CT) did not reveal enlarged lymphatic nodes or ureters obstruction (Figure 1 A-D). Due to radiologic suspicion of malignancy, tumor markers were assessed, with negative CEA and CA 19-9 results, and endocrine autonomy was excluded. Gastroscopy and colonoscopy were unremarkable. The patient was referred for surgery, where biopsy was performed because radical resection was not feasible.

Histological examination showed fragments of fibrous connective tissue with collagen fascicles, few scattered plasma cells (some IgG4 positive), macrophages, clusters of small lymphoid cells, and islets of isolated eosinophils, resorptive granulomas, but no tumor cells. The CK, B-catenin, and Alk-1 stains were negative. Additional tests for ASMA and ANCA were negative. The immunoglobulin concentration, including the IgG4 subclass, was normal. Idiopathic retroperitoneal fibrosis was diagnosed. The patient was started on pulse cyclophosphamide (600mg infusion every four weeks) and prednisone therapy, which was continued for one year. At the end of therapy, abdominal and pelvic MRI showed regression of the tumor size and fibrous complex to 68 x 52 x 56 mm. 17F-glucose PET-CT performed to assess the inflammation activity showed no increase in glucose metabolism within the tumor mass (SUVmax 2.61). In the posttreatment follow-up (at 6 months), the patient reported no pain, and the lesion was stable on the MRI (Figure 1E-F).

Ormond's disease is an IgG4-related disease (RD) characterized by a relatively low percentage of IgG4 plasma cells compared to other IgG4-RD. Typical symptoms include abdominal, flank, or groin pain, as well as other manifestations related to compression of adjacent structures. [1,2]. Cyclophosphamide in combination with glucocorticoids may be used with favorable outcomes [3], as confirmed by our case.

Radiologic imaging, including MRI, may be helpful for diagnosing and monitoring disease activity [4,5]. The radiologic findings in our case were not typical of Ormond’s disease, which contributed to the diagnostic difficulty. In atypical cases, such as the present one, imaging

studies cannot replace pathological examination [5]. We would like to emphasize that PET-CT may be useful for assessing disease activity. It may have adjunctive value in supporting the decision to continue or discontinue the ongoing therapy. Follow-up imaging is essential for disease monitoring.

In conclusion, Ormond's disease can present with various imaging appearances. An atypical radiologic presentation should not exclude Ormond's disease from the differential diagnosis. PET/CT may be used to assess disease activity and support therapeutic decision-making.

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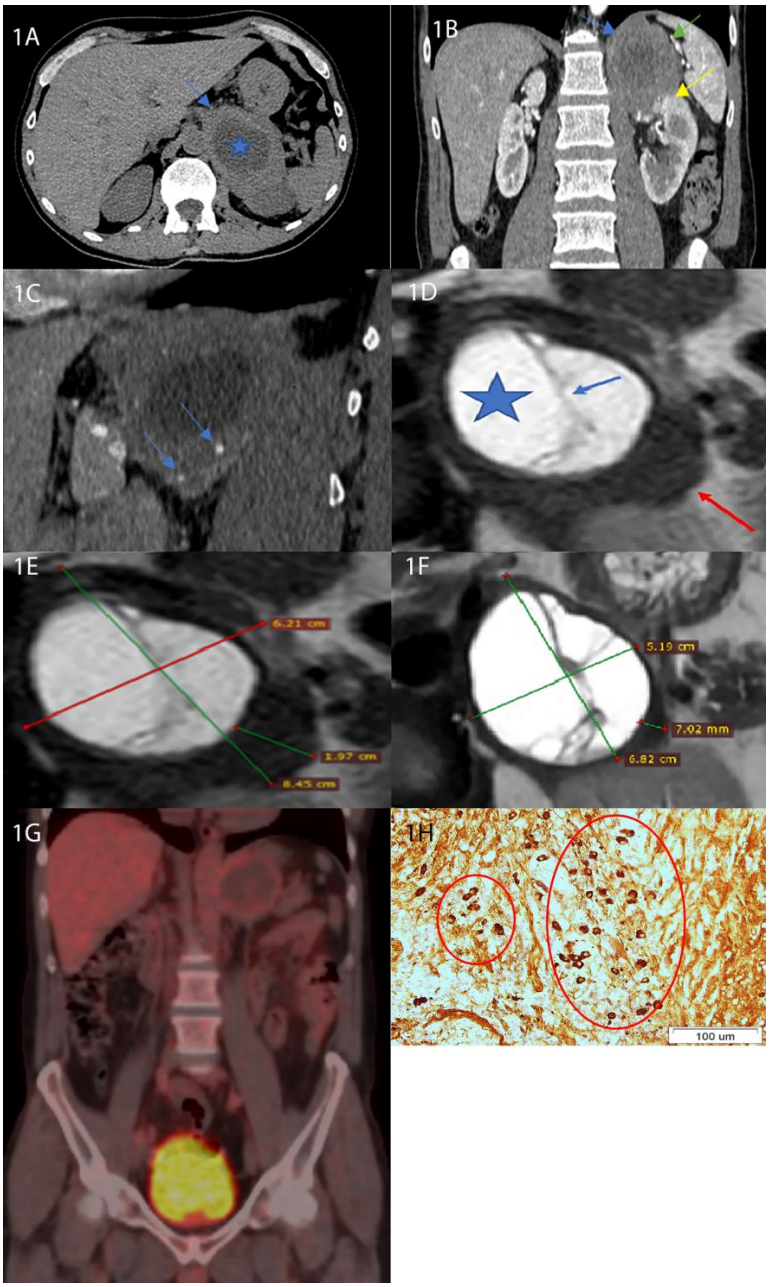

Figure 1 A – Large tumor in the left suprarenal region ( ) with thick wall and hypodense center ( ) most likely represents necrosis; **B** – the lesion in the left suprarenal region ( ) abuts the upper pole of the left kidney ( ) without evident invasion and slightly dislocates the spleen ( ); **C** – the tumor encases vessels without significant narrowing ( ); **D** – T2-W sequence. Initial MR study better visualizes thin septa ( ) in the hyperintense, fluid center ( ) of the thick, hypointense wall ( ) lesion in the left suprarenal region; **E** – T2-W sequence MR imaging. Lesion size and wall thickness before treatment; **F** – T2-W sequence MR imaging.

After six months of treatment, there is partial regression of lesion size and wall thickness; **G –** fluorine-18-fluorodeoxyglucose PET-CT, coronal reconstruction. No significant tracer uptake was observed in the left adrenal lesion on the PET-CT scan; **H** – inflammatory infiltrates of IgG4+ plasma cells. IgG4 staining. Magnification: 200x

Short title: An atypical presentation of retroperitoneal fibrosis: challenges